Communication and cognitive impairments and healthcare decision-making in MND: A narrative review

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Abstract

Rationale: Motor neurone disease (MND) is a neurodegenerative disease presenting with progressive weakness of voluntary muscles. For any condition, person-centred healthcare relies on the sharing of information and a mutual understanding of the person's needs and preferences. Decision-making in MND becomes more complex as there is no cure and a high prevalence of co-morbid communication and/or cognitive difficulties.

Objective: To identify the reported impact of communication and/or cognitive impairment on patient and carer involvement in healthcare decision-making in MND.

Methods: A review and synthesis of studies addressing issues of communication impairment and/or cognitive impairment in relation to decision-making focussed on MND was conducted. Articles were excluded if they were reviews, case studies, conference papers or commentaries. To be included studies needed to address issues of communication impairment or cognitive impairment specifically in relation to decision-making. Relevant data was extracted verbatim and subjected to content analysis to support the narrative summary.

Results: Seventy-six articles were identified and 35 articles screened. Six articles met inclusion criteria each describing examples of decision-making in MND. There was limited data related to communication and/or cognitive impairment and the impact these impairments may have on decision-making despite recognition that many people with MND may lose verbal communication or develop subtle cognitive impairments. The literature is primarily from the perspective of others.

Conclusion: This review highlights that the current body of literature exploring decision-making within the MND population presents us with extremely limited insights into the impact of communication and/or cognitive impairments on healthcare decision-making. Extant literature focuses on interventions (namely ventilation and gastrostomy), the broad process of decision-making, or cognitive assessment of decision-making ability. Whilst most studies acknowledge that deficits in communication or cognition impact the decision-making process, this issue is not the focus of any study.

Keywords: motor neurone disease, amyotrophic lateral sclerosis, decision-making, communication impairment, cognitive impairment

1. Introduction

Person-centred healthcare decision-making uses a shared approach and is considered an ideal treatment decision-making process [1]. Shared decision-making principles include eliciting and acknowledging patients' preferences for participation, patient choices regarding how decision-making processes will proceed, and respect and adherence of patient choices by their clinicians [1]. This approach involves the sharing of knowledge, values and preferences, by clinicians and patients, to deliberate together, so that decisions are congruent with patient values and preferences [2]. This process highlights the importance of clinicians and patients working together to facilitate patient autonomy [3]. Decision-making becomes more complex when there are no curative treatment options and when there is a high prevalence of communication difficulties and a lesser prevalence of cognitive difficulties within a context of a rapidly progressive condition, as seen in motor neurone disease (MND).

Motor neurone disease is the name given to a group of neurodegenerative diseases characterised by insidious onset muscle weakness with onset usually focal in the limb, bulbar or truncal musculature resulting in the loss of mobility and verbal communication [4]. Degeneration of both upper and lower motor neurones causes impaired function of limbs, communication, cognition, swallowing and breathing [5]. Disease course, site of onset and progression, as well as symptoms experienced, vary greatly. Although significant disease duration variability is reported, most patients die within a few years of symptom onset. Fifty percent of patients die within 30 months of symptom onset and approximately one-fifth survive between five and 10 years [6]. Approximately 80-90% of people

diagnosed will develop a motor speech impairment [7] and current literature estimates cognitive deficits are present in 25-50% of patients [8]. It is not clear from the literature what proportion of individuals with MND have communication impairment and co-occurring cognitive deficits; therefore throughout this paper the term communication and/or cognitive impairment will be used. Cognitive ability supports communication skills, consequently it is important to consider both. Because there is no means of halting or reversing the disease, understanding the impact of communication and/or cognitive impairment on healthcare decision-making is significant in this population, given that the clinical focus is symptom management and quality of life.

2. Background

Healthcare decision-making in MND is especially complex due to the nature of decisions required, variation in personal values and varying access to services [9]. Decision-making in MND can be influenced by access to specialist MND care, inclusion of family carers in decisions, and patients' personal reactions to their diagnosis and deterioration [10,11]. Carers make a significant contribution to decision-making in specialist care [12,13]. Deficits in communication, cognition and behaviour, as well as a patient's focus on the present, can be barriers to participation in decision-making [10,11]. A pro-active approach to interventions allows for timely medical procedures (e.g. feeding tubes, ventilatory support) and changes to the home environment. However, making treatment decisions before symptoms are obvious is challenging and further complicated in the presence of frontal cognitive deficits. Decision-making requires effective communication and cognitive skills to make appropriate choices and engage in all the above. Communication and cognitive changes are significant to patients and carers, and are prominent in international clinical guidelines which recommend ongoing consideration of people's needs to optimise communicative effectiveness, introduction of alternative communication devices, reassessment over the time course and consideration of decision-making capacity issues [4,14-16]. These issues however, remain under-researched.

2.1. Communication impairment associated with MND

Many people with MND will develop communication impairment. Dysarthria (difficulty speaking) is the most common communication impairment experienced in this population and will be experienced by 80-90% of people with MND [7]. Dysarthria is the first or predominant sign in the early stage of the disease for 25-30% of patients and is eight times more frequent than dysphagia (difficulty swallowing) [7]. Characterisation of dysarthria is based on site of involvement and the mix of lower and upper motor neurone features, with possible spastic, flaccid or mixed dysarthria occurring [7]. Speech in MND is typically characterised by the presence of hyper-nasality, distorted articulation of constants and vowels, strained-harsh or hoarse vocal quality and slow speech rate. However, consistent with the highly variable nature of the disease overall, dysarthria presentation also varies. Some patients may only experience an isolated dysphonia (difficulty with voice

production), usually related to poor respiratory support for speech output. Dysphonic patients find it hard to communicate in groups or noisy environments or when they are talking with someone with hearing impairment. Associated with the upper motor neurone component, (pseudobulbar) dysarthria is often accompanied by emotional lability (pseudobulbar affect). This causes exaggerated emotional outbursts of laughing or crying, which are difficult to control and often inappropriate to the circumstances. When prominent, these episodes are socially disabling and further impair communication, especially around emotive topics. Apraxia of speech (speech sound disorder) (AOS), another motor speech disorder, has been identified in individuals with MND, albeit uncommonly. In a study of AOS in a variety of neurodegenerative disorders, a small number of individuals were diagnosed with MND [17]. In these individuals, AOS was among the first symptoms of MND and was more prominent than the accompanying dysarthria.

2.2. Cognitive impairment associated with MND

Historically approached as a pure motor disorder, the cognitive changes associated with MND are becoming widely recognised and better understood [18], along with the recognition of behavioural and linguistic changes. The presence of cognitive impairment can be easily missed due to patients not reporting difficulties, general lack of awareness of cognitive deficits by people with MND, carers and clinicians, or impediments to neuropsychological testing such as speech deficits, or other motor problems such as difficulty writing. Assessment of cognitive function may not be given high clinical priority in the context of a rapidly progressing disease, with medical and functional treatment decisions taking priority [19,20]. Changes in cognitive function vary from subtle cognitive impairment to frank frontotemporal dementia (FTD) [8]. Recent studies of patients without overt dementia, drawn from specialist ALS clinics in the UK, showed that a quarter of patients met the criteria for behavioural variant frontotemporal lobar degeneration on formal testing [18,21]. Additionally, almost half of participants without cognitive impairment on formal cognitive screen presented with frontal dysfunction, behavioural change, impaired emotional processing and/or impaired decision-making [18,21]. Apathy was the most common behaviour dysfunction reported. Whilst apathy is pervasive, there is no clear link to specific MND phenotypes [16]. Other common deficits consistent with a previously reported MND cognitive profile were executive dysfunction, naming difficulties and impaired verbal fluency [8]. Assessment of cognitive function is vital in order to understand patients' cognitive capacity and to appropriately support their planning and decision-making [19,20].

2.3. Language impairment associated with MND

Language impairments in MND are an emerging area of MND research. Impaired spelling, confrontation naming, single word comprehension, verb processing and sentence grammar have been found in patients with executive function scores within normal limits, meaning that executive dysfunction was not a contributing factor to language deficits [8,21,22]. The results of these studies

provide evidence that language impairment is as prevalent as executive dysfunction in MND; however, the profiling of linguistic dysfunction is challenged by the association between language and executive functions, impaired speech and other motor deficits on language assessment, and compounded by the heterogeneity of presentations within the population. A small number of cases of aphasia in MND have been described in the literature with patients presenting with comprehension deficits, non-fluent aphasia, anomia and dysgraphia [23]. In fact, changes in language function should be considered an important characteristic of MND and are not necessarily explained by an overlap of ALS and FTD [16,23]. Other well described pragmatic features of communication dysfunction in MND include impaired social communication, impaired facial expression, emotional lability, and pseudobulbar affect, which all impact on interpersonal communication. The inability to express, or to read, social communication cues has the potential to negatively impact interpersonal relationships. In the context of severe physical and speech disability, familial and social relationships have an increased significance and correlation to quality of life [24]. In this review, motor speech impairment, linguistic and social communication difficulties are collectively referred to as communication impairment.

2.4. Impact of communication and/or cognitive impairment

Loss of or reduced effectiveness of communication prevents patients from participating in many activities, can lead to social isolation, and can reduce quality of life for both patients and carers [4,25]. Difficulty with expressive communication results in significant vulnerability and increases reliance on caregivers for communication support, including making healthcare decisions [25,26]. The loss of effective communication can result in intellectual and emotional isolation for carers [4]. Communication impairment is linked to increased medical error rates [26]. Preventable adverse events during acute hospital admissions are three times more likely to occur in patients with a communication impairment than in patients without communication difficulties [27]. Additionally, the failure to assess for cognitive deficits and decision-making capacity has significant healthcare implications for patients. Communication and/or cognitive difficulties may manifest as problems making decisions, managing finances, planning, and learning new tasks [4]. Motor impairments such as upper limb dysfunction make writing and the use of alternative communication devices problematic, resulting in additional difficulties engaging in healthcare decisions without support. Decision-making for people with communication impairment associated with motor impairment can be more easily circumnavigated with alternative communication devices and communication strategies, whereas decision-making for people with cognitive difficulties requires more structured facilitation.

There is limited understanding about the impact communication and/or cognitive impairment has on the decision-making process for people with MND and their carers. This is despite the high prevalence of these disorders in MND and numerous clinical guidelines recommending adjustments

to clinical care to account for patients' communication and/or cognitive difficulties. This review aims to synthesise current knowledge of the impact of communication and/or cognitive impairments on healthcare decision-making in MND.

3. Methods

Search strategy and assessment

Our review was based on a search conducted on electronic databases Medline, PsycINFO, Emcare and CINAHL for peer-reviewed research published in English from January 1998 to November 2018. Search terms used were either as MESH or subject heading terms: motor neuron* disease OR amyotrophic lateral sclerosis AND (decision making OR advance care plan OR advance directive) AND (communication disorder OR cognition disorder). The reference lists of reviewed articles were searched to identify any further relevant research, which identified three additional studies for review. This is depicted schematically in Figure 1 below.

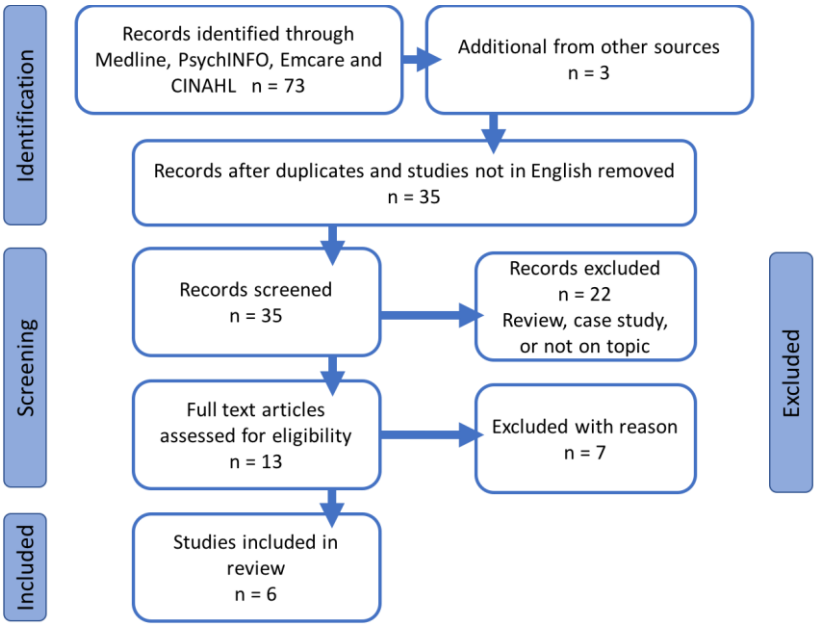

Figure 1: PRISMA flow diagram summarising the literature search.

To be eligible for inclusion in our review, studies need to address issues of communication impairment or cognitive impairment in relation to decision-making. The search included empirical studies and did not include reviews, case studies, conference papers or commentaries. All articles were identified by author CP and 20% of records screened were checked by MC to ensure they met eligibility for inclusion. Where appropriate, studies were evaluated using the Critical Appraisal Skills Program Qualitative Research Checklist (CASP) [28]. This tool assesses the quality of the study using criteria to measure data collection and analysis, recruitment, ethical issues and overall

contribution of the study to the body of knowledge. The checklist confirmed four of five studies evidenced valid results with appropriate methodology and clear analysis methods (one study was not appropriate to assess using the CASP as it was published as a short Clinical/Scientific Note) (see Table 1 for summary). The initial intention had been to employ thematic analysis to synthesise the results however given the limited data available, relevant data was extracted verbatim from each paper and subjected to content analysis to support the narrative summary. As this review did not involve human subjects, ethical approval was not required.

Table 1. Methodological rigor of included studies (CASP Checklist[28])

Study (first author last name)	Was there a clear statement of the aims of the research?	Is a qualitative methodology appropriate?	Was the research design appropriate to address the aims of the research?	Was the recruitment strategy appropriate to the aims of the research?	Was the data collected in a way that addressed the research issue?	Has the relationship between researcher and participants been adequately considered?	Have ethical issues been taken into consideration?	Was the data analysis sufficiently rigorous?	Is there a clear statement of findings?
Hogden 2012	Yes	Yes	Yes	Yes	Yes	No	Yes	Yes	Yes
Hogden 2013	Yes	Yes	Yes	Yes	Yes	No	Yes	Yes	Yes
Lemoignan	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes/Can't tell	Yes
McKelvey	Yes	Yes	Yes	Yes	Yes	No	No	Yes	Yes
Preston	Yes	Yes	Can't tell	Yes	Can't tell	No	No	No	Can't tell
* Bohm 2016 not appropriate for quality assessment due to being published as a short Clinical/Scientific Notes									

4. Results

Seventy-six articles were identified for screening; 73 from database extraction and three additional articles identified from reference lists. Thirty-five articles were screened once duplications or studies not available in English were removed. A further 22 articles were excluded as they were a review, conference paper, case study or opinion piece, or based on title or abstract information. Thirteen articles were reviewed, with seven *excluded with reason* as they did not meet the inclusion criteria. Of the seven excluded articles, almost half discussed determinants or attitudes to shortening or prolonging life, or end-of-life care but did not discuss the impact of communication and/or cognitive impairment on patient attitude, autonomy or involvement in treatment or end-of-life care decision-making. The remaining articles either described the types of augmentative and alternative communication (AAC) that may be used in MND or described clinical assessment of cognitive function.

Six articles met the criteria for this review. Five employed cross sectional, qualitative methodology (four employing thematic analysis, one phenomenology), two being retrospective (i.e. interviewing bereaved carers), and one study employed quantitative methodology (See Table 2). The papers within and of themselves provide rich and illustrated examples of decision-making in MND. However, throughout these studies there was very limited data related specifically to communication and/or cognitive impairment and the impact these impairments may have on decision-making. The literature was primarily from the perspective of others: three of the six studies interview carers, one study interviewed healthcare professionals, one study interviewed people with MND and their caregiver, and one study assessed with people with MND.

The loss of verbal or written communication and its impact is mentioned in three studies. Bereaved carers report that advance care planning documents would best be done when people can talk or write so they are fully able to express their feelings [29]. “Nudging a big button, a very slow word processor, is not the same as talking and I think … if we had done it [earlier, the patient] would have been able to express probably more forcefully her feelings" (p133). Carers felt their role to ‘promote the patient’s voice’ was more challenging when patients lost their ability to speak [12]. This impacted the carer’s ability to support the patient’s decision-making style and required them to facilitate communication between the patient and healthcare professionals. In this study, six of the eight carers interviewed also assumed communication responsibilities such as dealing with services, when their family members lost verbal communication, increasing reliance on family members [12]. The value of AAC to support and to enable engagement in healthcare situations, for patients who were no longer verbal communicators was evident in McKelvey et al. (2012), “we still made decisions together (but) she would write instead of talk” (p238) [30].

The issue of cognitive impairment and its impact on decision-making is discussed in three studies. In a large study of 169 patients from a specialised German ALS clinic, analysis found no association with patients' decisions regarding the use or decline of PEG, NIV and invasive ventilation or hypothetical hastened death, and cognitive impairment [31]. Contrastingly, two qualitative studies interviewing carers and healthcare professionals both reported cognitive impairment impacted patients' healthcare decision-making [12,32]. Carers reported it was more challenging to advocate for patients, and they needed to provide additional emotional support to facilitate patient decision-making, when cognitive changes had occurred [12]. Healthcare professionals identified an impact on the timing and quality of decisions, resulting in patients delaying decisions or making choices that may risk the well-being of carers [32]. They also acknowledged that subtle cognitive impairment was difficult to clinically identify. For example, "They do have some sort of cognitive impairment which is not enough to be dementia, but they're not functioning as they used to, and I'm sure it is impacting in some way" (p.694) [32]. The differing results in these studies may reflect the methodological restriction of standardised questionnaires in obtaining in-depth information from participants.

The one study interviewing people with ALS reported salient information regarding the high value participants placed on communication. All participants identified communication as the most important factor in deciding about respiratory failure treatment options [33]. "...As long as I can properly communicate with my voice, my eyes or a machine or whatever, I want to have a respirator... But as soon as I can no longer communicate, that's it! I don't want anything else to be done" (p210). Decision-making autonomy, being involved and respected in the process and "wanting to have the final say" (p210) was highly important for all participants [33].

Table 2. General information and summary of included studies

Study	Methodology	Sample	Primary focus of study	Findings
Medical decisions are independent of cognitive impairment in amyotrophic lateral sclerosis. Bohm et al. 2016	Quantitative analysis - logistic regression analyses	n=169 patients recruited from a German specialist outpatient clinic.	To identify the possible association between cognitive and behavioural impairment and decision-making in ALS.	Reported as a Clinical/Scientific note therefore findings are brief. - Participants were screened with the Edinburg Cognitive and Behavioural ALS Screen (ECAS), with 140 of participants' caregivers providing information regarding behavioural change. - Participants responded to standardised questionnaires regarding their decisions about percutaneous endoscopic gastrostomy (PEG), non-invasive ventilation (NIV) and invasive ventilation (IV). - Deficits in at least one cognitive domain was present in 55% of participants. - Caregivers reported apathy as the most commonly present behavioural change (15%) - Logistic regression analysis showed than neither cognitive impairment or behavioural changes were associated with participants' decisions regarding use or decline of PEG, NIV or IV, hypothetical ideation to turn off treatment, or wish for hastened death (all $p > 0.05$).
Engaging in patient decision-making in multidisciplinary care for amyotrophic lateral sclerosis: the views of health professionals. Hogden et al. 2012	Qualitative thematic analysis Cross sectional	n=32 healthcare professionals from two specialised ALS clinics (one metropolitan, one regional) in NSW, Australia.	To identify factors influencing patient decision-making from the perspectives of healthcare professionals.	Healthcare professionals identified barriers and facilitators to decision-making - Barriers included; patient's acceptance of the diagnosis, types of information patients sourced, and patient-carer relationship. - Facilitators included; collaborative multidisciplinary team working, effective communication systems, and evidenced-based clinical information. - Cognitive and behavioural changes impacted quality and timing of decisions although patients retained decision-making capacity despite subtle cognitive impairment. - Respondents supported the introduction of routine screening for cognitive and behavioural changes.
What are the roles of carers in decision-making for amyotrophic lateral sclerosis multidisciplinary care? Hogden et al. 2013	Qualitative thematic analysis Cross sectional	n=8 carers of ALS patients from two specialised ALS clinics (one metropolitan, one regional) in NSW, Australia.	To identify carer roles, and determine facilitators and barriers to carer participation in decision-making for ALS multidisciplinary care.	- Carers made significant contributions to ALS decision-making. - Four common carer roles identified were; promoting the patient voice, promoting patient health literacy, provision of emotional support, and logistical assistance. - Facilitators and barriers to carer participation in decision-making were identified. - Changes to patient communication and cognition was identified as a barrier to decision-making.

Amyotrophic lateral sclerosis and assisted ventilation: how patients decide. Lemoignan et al. 2010	Qualitative phenomenology Cross sectional	n=9 people with MND and caregivers. Maximum variation sampling. Recruited from a Canadian ALS Clinic.	To better understand the experience of decision-making about assisted ventilation for ALS patients.	- Six main themes were reported from the interviews; the meaning of the intervention, the importance of context, the importance of values, the effect of fears, the need for information, and, adaptation to or acceptance of the intervention. - Within the "importance of values" theme, participants emphasised the value of communication, and the ability to communicate was identified as the most important factor in deciding about treatment options. This influenced some participants' choice of intervention. - Participants reported valuing decisional autonomy, and being involved and respected in the decision-making process.
Communication styles of persons with ALS as recounted by surviving partners. McKelvey et al. 2012	Qualitative thematic analysis Retrospective Cross sectional	n=7 (6 bereaved spouses, 1 bereaved daughter) whose family members had used high- or low-tech alternative and augmentative communication strategies. Convenience sampling. Recruitment source not stated.	To describe the communication patterns of individuals with ALS over time as disease progressed, as reported from the surviving spouses' perspectives.	- Four primary themes were identified; communication styles, augmentative and alternative communication (AAC) use, decision-making, and lifestyle changes. - Loss of spoken communication was devastating to their partner, and impacted public and social participation. - Healthcare decisions were always made with input from their partners with ALS. - AAC devices were described as essential for individuals with ALS to continue decision-making and to facilitate participation in medical decision-making.
The Preferred Priorities for Care (PPC) document in motor neurone disease: views of bereaved relatives and carers. Preston et al. 2012	Qualitative thematic analysis Retrospective Cross sectional	n=11 bereaved relatives or carers. Recruited from MND Care & Research Centre in N.W.England.	This study examined bereaved relatives' experiences of using the PPC (a patient-held record of advance care plans). Research explored relatives' perceptions about its impact on end-of-life care.	- Four main themes were identified specific to the PPC; completion, document availability, important and influence on end-of-life experience, and limitations. - Several participants felt the PPC best completed whilst patients were able to write or talk - PPC contributed positively to end-of-life experience by providing a sense of relief and peace of mind, however the document had little practical impact on end-of-life experience due to other people's awareness of patient preferences irrespective of the physical document. - Appropriate timing of advance care planning is highly individual however is more effectively achieved whilst the patient retains the ability to communicate effectively - Significant role of the PPC in raising awareness of patient preferences amongst healthcare professionals for a patient group where effective verbal communication is often lost before the end of life.

5. Discussion

There is a paucity of research specifically addressing the impact of communication and/or cognitive deficits on decision-making for people with MND and their carers. There are a number of caveats to the findings of those studies. The majority of the research uses cross-sectional methodology; however, single interviews can sometimes fail to capture the implications of ongoing change. Except for Bohm et al. (2016) and Hogden et al. (2012) the studies had small sample sizes. Much of the work obtains data from bereaved carers and whilst the experiences and perspectives of carers are particularly important because carers typically serve as the primary support, there remains a gap in the literature in regard to the direct exploration of the experiences of people with MND, particularly as the disease progresses.

Although there is a significant and growing body of literature on decision-making within the MND population, the impact of communication and/or cognitive impairments does not emerge with any clarity. Instead, the broader extant literature focuses on particular interventions (namely ventilation and gastrostomy) [33-39]; the broad process of decision-making (from the perspective of the patient, carer or healthcare professional) [10,12,13,32,40-42]; or cognitive assessment of decision-making ability [43,44]. However, this information is rarely extrapolated to consider patients' personal decision-making. Almost all studies acknowledge that communication and cognition are important for decision-making capacity and for patient autonomy. Even though most studies acknowledged the need to facilitate decision-making and support communication and cognitive deficits, only two papers provided specific strategies. Martin et al. (2015) provide extensive strategies to support people with ALS and their caregivers in their decision-making (see paper for detail) [37]. Strategies suggested by people with severe speech disabilities, including people with ALS (but not limited by cognitive impairments) for use with physicians in the context of primary care appointments (but useful in any medical or health encounter) include: allow more time for communication; tell patients if they're not understood; use appropriate language and non-verbal communication; and speak with the patient not just the companion [45]. Finally, decision-making models often fail to acknowledge communication and/or cognitive impairment even though they endeavour to capture the complexities of decision-making for patients, families, health professionals, researchers and policymakers [9].

The high importance of communication to participants is apparent in the broader literature, as well as the articles included in this review. It is described as the *most important thing*, or the loss *most felt*. In a study describing the communication patterns of individuals with ALS, a bereaved carer reports "that was probably her biggest hurt. She couldn't talk." (p235) [30]. Participants linked the ability to communicate to their values of decisional autonomy, and the importance of being involved and respected in the decision-making process [33]. Communication needs to be prominent in decision-making literature and in decision-making models due to its high value to patients and family members, to facilitate patient autonomy, and person-centred care.

Timeliness of decision-making is frequently raised in the broader MND literature. A discord is evident between people with MND and healthcare professionals, or between people with MND and their family carers, with healthcare professionals and family carers often advocating for early or pre-emptive decisions while people with MND, either due to denial or wishing to focus on the present, choosing to postpone decisions [10,36,47]. The importance of early decision-making is often justified due to the inevitable deterioration in speech function and possible development of cognitive deficits [20,29]. Early discussions with patients and carers are recommended in international clinical guidelines for MND, in order to establish advance care plans or because timing is crucial to some intervention decisions, such as gastrostomy, which has a defined window of opportunity [4,14,15]. Early decision-making has further clinical implications, for example, the early adoption of AAC promotes successful implementation [48] which is why the adoption of a pro-active decision-making style, even if not in keeping with pre-morbid decision-making approaches is encouraged [10,48]. A similar tension of timing is evident when participants were asked about information counselling. Patients reported valuing information, but not all wanted it before it was needed to make decisions which was often in contrast to family members, who report that information was a means to feel empowered and in control, and therefore wanted information at the onset of the disease [12,13,32,33]. The literature supports decision-making to be largely individual, and delivered at the client's pace, with generic tools or algorithms described as inappropriate [8,9,13,49].

The studies included in this review, as well as the broader literature, highlight related points that affect the decision-making context for people with MND and their carers. People with MND experiencing communication and/or cognitive impairments rely on both carers and communication devices (either low-tech or high-tech) to facilitate communicative interactions such as decision-making. However, it must be acknowledged that not all patients will have family to rely on, or who are able to be present for clinic appointments or involved in information sharing and decision-making due to their own medical, financial or employment issues. This significantly increases the vulnerability of this group of patients. Exposure to unnecessary vulnerability is experienced by some patients when accessing AAC. The value of AAC to carers has been identified in reducing carer burden, facilitating greater rewards and social closeness, and reducing difficulty of providing care for people with ALS using AAC [50,51]. AAC has a critical role in enabling patient autonomy, however challenges to accessing AAC are reported and is unacceptable. People with ALS report that residing in a rural location, the logistics of navigating funding support for AAC and the knowledge-base of their healthcare professionals all impacted AAC access and obtaining a communication device in a timely manner [30,46,48]. This is despite evidence that early adoption promotes successful implementation [48]; "The insurance just finally paid for his talking machine, shortly after he died, I finally got the check" (p237). A further barrier to access is identified by a bereaved carer who describes needing to set up AAC during an inpatient admission; "it's unfortunate that most [hospitals] don't know how to set up his speech equipment" (p236) [30]. These issues clearly identify a need for education and training for various professionals involved in the provision and use of AAC for people with MND, and whilst AAC use may be uncommon,

healthcare professionals would always have access to training if their patients' equipment related to the provision of medical or life survival interventions.

This review provides further evidence that studies addressing the association between, or the impact of, cognitive impairment and decision-making are scarce and currently undetermined [9,31]. However, the need to screen for cognitive changes, and the impact of cognitive changes on decision-making, as well the impact for carers, comes through strongly in the studies included in this review. This issue is reflected in international guidelines and other research, and it is clear that changes in cognition have implications for communication, decision-making and care provision with recommendations to appropriately screen and train both healthcare professionals and carers [4,15,44]. Cognitive screening strengthens clinicians' ability to recognise and address barriers to communication and decision-making, thereby enabling patient autonomy [43]. Within the context of clinical decision-making, it appears that cognitive impairment is discussed more frequently than communication impairment in the broader literature. This may reflect that communication impairment associated with motor impairment can be more easily circumnavigated with AAC and communication strategies, whereas decision-making for people with cognitive difficulties requires more structured facilitation. It may also reflect medicolegal issues of mental capacity and capacity to consent in decisions. Health professionals identified both insufficient health-service funding, and a lack of a systematic cognitive assessment in their setting, as potential barriers to patient-centred decision-making [32]. Historically, cognitive dysfunction in MND has not been well recognised resulting in a need for upskilling of healthcare professionals in the identification, screening and assessment of cognitive disorders.

6. Conclusion and Future Directions

This review highlights that the current body of literature exploring decision-making within the MND population presents us with very limited insights into the impact of communication and/or cognitive impairments on healthcare decision-making. From the broader literature, it is clear that people with MND acutely feel the loss of communication abilities, that they require personalised decision-making influenced by their acceptance of diagnosis and their decision-making style, that they rely on carers and communication devices to support their decision-making, and they require support and facilitation from healthcare professionals. We have identified factors that affect decision-making when communication and/or cognitive impairments exist such as a barrier to the full expression of feelings, increased reliance on carers, and the impact of timing on the quality of decisions.

Until the impact of communication and/or cognitive impairments on decision-making is fully understood, people with MND and their carers will not be completely supported. Future research should focus on understanding the factors that support communication and decision-making effectiveness for people living with MND over the course of the disease. This is vital for patient-centred care and to support families and carers. A longitudinal study interviewing people with

MND and their carers across the stages of this disease would provide insights to the real-time experience of making and living with decisions as time and the disabilities of MND progress.

Acknowledgements

Ms Paynter acknowledges the financial support of a MNDRIA ‘top up’ grant.

Declaration of interest

The authors report no conflict of interest.

Financial disclosure statement

Ms Paynter is funded by a joint NHMRC/MNDRIA postgraduate scholarship.
A/Prof Cruice reports no relevant disclosures.
Dr Mathers reports no relevant disclosures.
Dr Gregory reports no relevant disclosures.
A/Prof Vogel is funded by National Health and Medical Research Council, Australia Dementia Fellowship (#1135683).

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